

Recurrent venous obstruction caused by sarcoidosis

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Case Report

A 45-year-old married, smoking Asian male was referred to the chest clinic in 1988 on account of an abnormal chest X-ray, having been seen and treated by an ophthalmologist for anterior uveitis, typical of sarcoidosis with pars planitis and vitreous snowballs. His only symptoms were dizziness and a feeling of 'blood rushing to his face' on coughing. On examination, there was superior vena caval obstruction, but no associated cervical lymphadenopathy or skin lesions. Investigation included a normal bronchoscopy and bronchoalveolar lavage (82% macrophages, 13% lymphocytes), bronchial biopsies deemed insufficient for diagnosis, a weakly positive mantoux at a concentration of 1:100, a normal serum angiotension converting enzyme (ACE) level and corrected serum calcium. A low total lymphocyte count was present $0.4\text{--}0.6 \times 10^9 \text{ l}^{-1}$ and the erythrocyte sedimentation rate was 49 mm min^{-1} . Pulmonary physiology revealed a mixed obstructive, restrictive defect with a forced expiratory volume in 1s (FEV₁) 1.46 l (43% predicted), forced vital capacity (FVC) 2.09 l (51% predicted), total lung capacity (TLC) helium 4.34 l (76% predicted) and a transfer factor of $22 \text{ ml min}^{-1} \text{ mmHg}^{-1}$ (80% predicted). A postero-anterior radiograph of his chest showed a widened superior mediastinum with bilateral paratracheal, and hilar adenopathy. At this stage, the differential diagnosis was one of sarcoidosis, lymphoma, tuberculosis, a mediastinal tumour or mediastinal fibrosis, and thus further investigation was undertaken. A computerized tomographic (CT) scan of his thorax confirmed the lymphadenopathy seen on the plain film, with compression of the left brachiocephalic vein and superior vena cava. A CT-guided biopsy of the subcarinal lymph node mass was performed, obtaining both solid material and straw-coloured fluid, both of which were sent for cytological examination. This specimen contained macrophages, lymphocytes, multinucleated macrophages and epithelioid cells; suggestive but not diagnostic of a granulomatous process. A Kveim biopsy contained non-caseating granulomata leading to a diagnosis of sarcoidosis.

Due to recurrence of his uveitis and persisting symptoms of cough and 'pressure in his neck', he was treated with oral

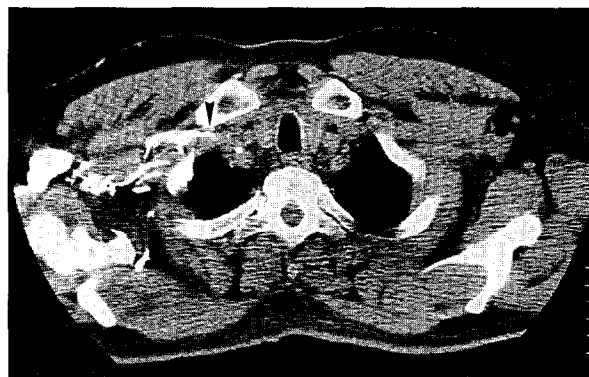


PLATE 1. Computerized tomographic (CT) scan of thorax, post contrast, demonstrating external compression, of the right subclavian vein at the point of entry (arrowhead) into the thorax.

corticosteroids, initially 30 mg prednisolone daily, reduced over a period of 20 months (April 1989–November 1991). This resulted in a prompt resolution of his clinical symptoms and also a markedly improved pulmonary physiology (FEV₁ 1.96 l, FVC 2.98 l, TLC helium 4.66 l and a transfer factor of $29 \text{ ml min}^{-1} \text{ mmHg}^{-1}$ (80% predicted). During 1991, a repeat CT scan of the thorax showed a reduction in the size of the lymph nodes; in addition, some now contained foci of calcification. The patency of the intrathoracic vessels was assessed using ultrasound, as the patient refused to have intravenous contrast. This indicated free flow in the subclavian veins. A further CT scan in 1993 was unchanged.

He presented earlier than planned in 1995 with a 2-day history of swelling of his right arm. On examination, his right arm was grossly swollen with normal arterial pulses and no axillary or cervical lymphadenopathy. The serum ACE level measured on this occasion was increased for the first time (132 IU l^{-1}). Pulmonary function had deteriorated again compared with 1991, with FEV₁ 1.66 l and FVC 2.41 l recorded. Right upper limb venography indicated complete occlusion of the right subclavian vein secondary to external compression, with associated thrombus within the distal vein. A CT scan of the thorax pre and post contrast demonstrated multiple nodes causing external compression, and occlusion of the right subclavian vein (Plate 1). The patient declined further investigation. He was treated with anticoagulants and oral corticosteroids (30 mg prednisolone daily), and his symptoms rapidly settled accompanied by a fall in serum ACE to 55 IU l^{-1} and

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improvement in his pulmonary physiology; FEV₁ 1.96 l and FVC 3.18 l recorded.

Discussion

The first descriptions of disease currently recognized as sarcoidosis were documented at the end of the 19th century and concerned skin eruptions (1). Since that time, it has been realized that it is a systemic disease characterized histologically by non-caseating granulomata. An aetiological agent eludes characterization. A number of recognizable patterns of disease at presentation are documented including Lofgren's syndrome (2). Lymph node enlargement is frequently found in sarcoidosis, occurring in up to 90% of patients during the course of their disease. The most common pattern of mediastinal adenopathy is bilateral involvement of the bronchopulmonary, right paratracheal and aortopulmonary nodes, but any combination can occur (3). Despite this, the incidence of obstruction of the intrathoracic vasculature is low. To the authors' knowledge, only four reports of superior vena caval (4–7) and one of innominate vein (8) obstruction due to sarcoid are described in the literature. No cases of subclavian vein obstruction have been reported previously. Obstruction of vessels can be caused by two mechanisms: involvement of the vessel wall with granulomata or extrinsic compression by enlarged lymph nodes. In the present case, the obstruction has been demonstrated radiographically (Plate 1) to be due to external compression by intrathoracic lymphadenopathy; however, concurrent microscopic involvement of the vessel wall could not be excluded.

The diagnosis of sarcoidosis in this case is supported by many factors: the initial presentation of uveitis and lymphadenopathy, abnormal pulmonary physiology which improved with oral corticosteroids, the positive Kveim, the

intrathoracic biopsy material obtained, and the raised serum ACE during the current recrudescence. The subsequent course of the illness and response to therapy would not be typical for any other granulomatous disease.

To the authors' knowledge, this case is the first to present on two separate occasions with intraathoracic venous obstruction, the second with an increase in activity of disease. At present, the symptoms have resolved with anticoagulation and corticosteroid therapy.

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Bilateral endobronchial metastases due to a chondroblastic osteosarcoma

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Introduction

Endobronchial metastases from non-pulmonary neoplasms are rare. When they occur, the primary site of the

tumour is most commonly breast, colon or kidney (1), although other sites have been reported, including sarcoma (2). The present case report describes a case in which bilateral metastases secondary to a chondroblastic osteosarcoma of the spine presented acutely with dyspnoea, and removal of these lesions resulted in great symptomatic improvement. However, the patient's subsequent rapid deterioration and death highlights that the prognosis in such cases is poor.

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